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Case Report

Successful percutaneous management of Lutembacher syndrome

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ABSTRACT

Background: The surgical management of Lutembacher syndrome is straight forward but percutaneous management, though technically demanding, is always desirable.
Methods: A 17 year old unmarried female presented with severe Mitral stenosis and a 19 mm almost circular Ostium secundum ASD with moderate pulmonary artery hypertension and dilated right sided chambers. She was managed in a staged manner. Percutaneous trans mitral commissurotomy (PTMC) was done first, using a 26 mm Inoue balloon catheter set, and after 48 h, ASD was closed with a 20 mm Cocoon Septal Occluder.
Results: The mitral valve area increased after PTMC from 0.8 cm^2 to 2.1 cm^2 and Q_P/Q_S decreased from 4.9 to 2. ASD was successfully closed under echocardiographic and fluoroscopic guidance.
Conclusion: Percutaneous management of the Lutembacher syndrome (PTMC and ASD device closure) is an effective and low risk procedure and avoids considerable morbidity and mental trauma for the patients.

1. Introduction

Role of surgical management in Lutembacher syndrome is well established. As this syndrome is more common in young females,[1] percutaneous management is always desirable. Here we present one such case of a 17 yr old female who was successfully managed by percutaneous technique.

2. Case

2.1. Patient details

A 17 year old unmarried female presented with a chief complaint of palpitation on exertion for last 4 years. On examination patient had RV type apex, S2 was fixed and widely

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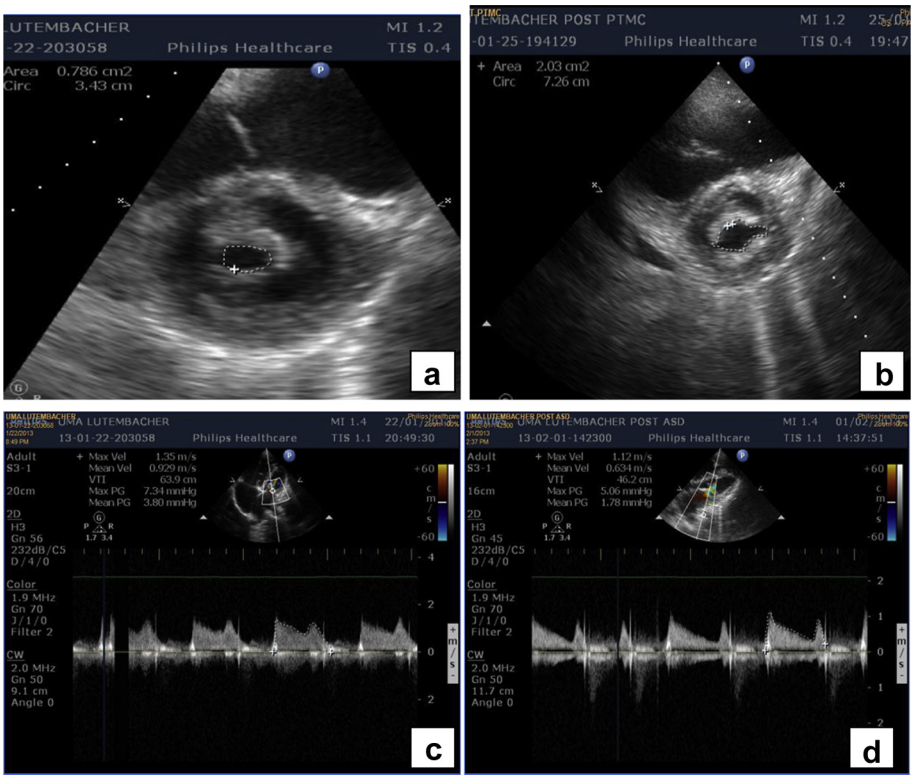

Fig. 1 – a and b) Parasternal short axis on TTE pre and post PTMC respectively. (c and d) CW Doppler across MV pre and post PTMC respectively.

split with loud P2 and grade 3 ejection systolic murmur in pulmonary area. On investigations, EKG showed incomplete RBBB, right axis deviation (120°), poor R wave progression in the precordial leads, diffuse T wave inversion and r/s in V6 <1 all s/o right sided volume or pressure overload and chest X-Ray showed cardiomegaly with RV apex (carried below diaphragm) and RA and MPA enlargement with increased pulmonary vascular markings s/o PAH but with no e/o PVH (no cephalization). Diagnosis of Lutembacher syndrome was confirmed by Transthoracic 2D-echocardiography (TTE), that showed mitral valve area (MVA) of 0.8 cm^2 by planimetry, with pliable valves (Wilkins score[2] of 5) (Fig. 1a and c) and by

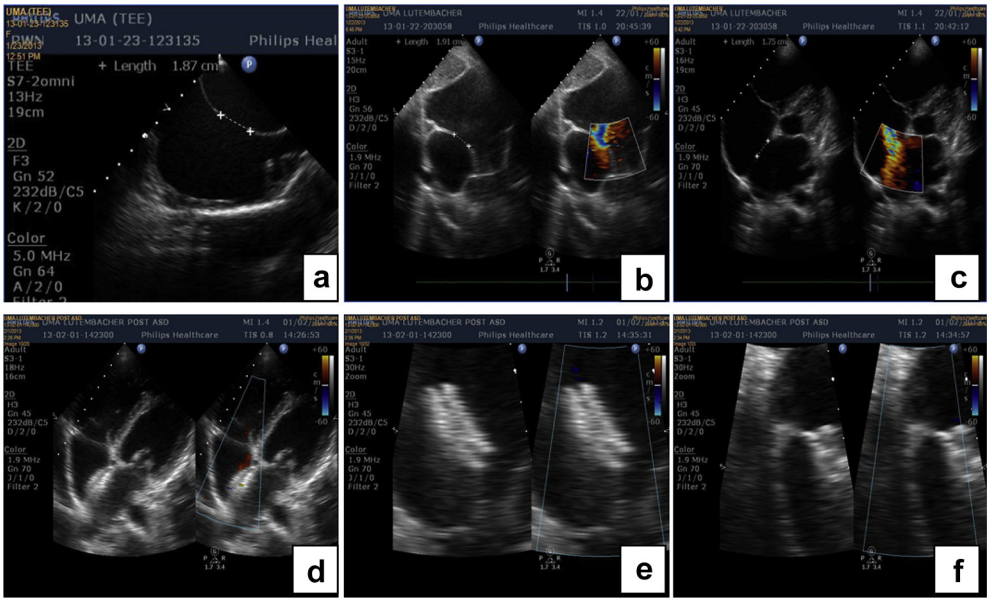

Fig. 2 – (a, b and c) OS-ASD in bicaval view on TEE, modified subcostal bicaval view and apical 4 chamber view on TTE. (d) The ASD device in apical 4 chamber view and (e and f) Subcostal views showing device holding SVC and IVC rims tightly respectively.

Transesophageal echocardiography (TEE) that showed 1.9 cm OS-ASD with adequate rims (Fig. 2a, b and c).

2.2. *Technique*

Oxymetry and pressure recordings were done twice, once before Percutaneous Trans Mitral Commissurotomy (PTMC) and second, 48 h after PTMC (Table 1). PTMC was done using a 26 mm Inoue balloon catheter set (Toray International America Inc. Houston, TX, USA) under fluoroscopy. We entered left ventricle (LV) with the help of the stylet after positioning the Inoue balloon in the left atrium (LA). ASD was closed using a 20 mm Cocoon septal occluder (Vascular Innovations Co., Ltd) (Fig. 2d, e and f), 48 hrs[4] after the PTMC, in a staged manner after confirming the optimal results of PTMC (Fig. 1b and d), and after determining the residual impact of OS-ASD on the hemodynamics of the heart after the PTMC (Table 1).

3. Discussion

PTMC in Lutembacher syndrome is associated with lower complications as it does not require septal puncture. Sometimes ASD is very posterior and to facilitate LV entry we may need to take separate anterior septal puncture. Even when complications do occur, they are hemodynamically benign as the blood preferentially flows to RA, which is a relatively low pressure chamber. During PTMC, we entered the LV without any difficulty. This was probably due to good anterior and antero-inferior margins of ASD that provided adequate support for the positioning of the balloon. If there is a difficulty[5] in entering the LV then one can use several techniques mentioned in the literature.[6–8]

Oxymetry, after PTMC, showed that step up at the RA level decreased from 31% to 7% and A-V O_2 difference decreased from 39 ml/min/m2 to 22 ml/min/m2 (suggestive of increased cardiac output). In view of successful PTMC and residual $Q_p/Q_s = 2$[9] (Table 1 and Fig. 1) after PTMC, it was decided to close the ASD. It is important to assess the success of the PTMC because if suboptimal result (redo BMV is not possible transseptally due to ASD device) or any complication like more than moderate MR occurs, that increases the likelihood of surgery in the near future, then one should avoid ASD device closure and should refer the patient for surgery. So routinely the ASD closure is deferred by 2–3 days for proper assessment of the outcomes of PTMC. Sometimes ASD is hemodynamically significant only in the presence of MS and after successful PTMC it may not require any further management.

Table 1 – Pre and post PTMC, Flow and resistance parameters.

Parameters	Pre PTMC HR –74 bpm VO_2 –120[3] ml/min/m^2	Post PTMC (after 48 hrs) HR –74 bpm VO_2 –120[3] ml/min/m^2
Q_p L/min	9.2	6.7
Q_s L/min	1.9	3.34
PVRI WU/m^2	1.0	0.45
SVRI WU/m^2	42	30
Q_p/Q_s	4.9	2

HR – heart rate; VO_2 – oxygen consumption; Q_p – pulmonary blood flow; Q_s – systemic blood flow; PVRI – pulmonary vascular resistance index; SVRI – systemic vascular resistance index.

4. Conclusion

Though the experience with percutaneous management of Lutembacher syndrome is restricted to few case reports[10–14] but with proper patient selection and detailed hemodynamic assessment, Lutembacher syndrome can be successfully managed percutaneously and the morbidity and mortality associated with surgery can be avoided.

Conflicts of interest

All authors have none to declare.

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